e- ISSN 0976-0342
Print ISSN 2229-7456

International Journal of Pharmacy & Therapeutics

Journal homepage: www.ijptjournal.com

Review article

LEONTIASIS OSSEA SYNDROME: A REVIEW

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ABSTRACT

Leontiasis ossea, also known as leontiasis, lion face or Lion Face Syndrome, is a rare medical condition used to describe a number of conditions that result in the affected patient's face resembling that of a lion. It is characterized by an overgrowth of the facial and cranial bones. It is most frequently associated with craniofacial fibrous dysplasia, other conditions may mimic leontiasis ossea including Paget's disease, fibrous dysplasia, hyperparathyroidism and renal osteodystrophy. The medical term leontiasis ossea is reported to be first coined in 1864 by the famed Virchow. Lion face syndrome is quite rare and involves extra growth of the bones of the face, particularly the maxilla or upper jaw. Growth in this area narrows the nasal opening, modifies the mouth, and can even press on the eye orbits and optic nerve, compromising vision . Less common form affects all the cranial bones as well as those of the face. Suffering from this form person loses all senses one by one and it leads to death from cerebral pressure. The overgrowth can lead to breathing and eating challenges. It can occur at any age. The symptoms include cluster headache, fibrous dysplasis, nasal obstruction, mutation in the POFUT1 gene and TWIST2 gene. There is no treatment other than exposing the overgrown bone, and chipping away pieces, or excising entirely where possible.

Key Words:- Leontiasis ossea, fibrous dysplasia, paget's disease, hyperparathyroidism, osteodystrophy.

Access this article online

Home page: http://ijptjournal.com/

DOI: http://dx.doi.org/10.21276/ijpt.2019.10.3.6

Quick Response code

Received:03.04.19 **Revised**:14.04.19 **Accepted**:25.04.19

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INTRODUCTION

Leontiasis ossea is a rare medical condition which is characterised by an overgrowth of the facial and cranial bones secondary to chronic renal failure and secondary hyperparathyroidism, renal osteodystrophy, paget's disease and fibrous dysplasia characterised by craniofacial, ribs, long bones and spine deformation (Dehghan P, Ali Z, 2015; Harut H, Aron B, 2014;) .Virchow, in 1864 first described leontiasis in a patient as the thickening of facial bones (Harut H, Aron B, 2014; Lee VS *et al*., 1996). Usually, this condition involves one or both maxillary bones which progressively increase in size and expand into the orbital, mouth, nose and adjacent sinuses (Haroyan H *et al*., 2015; Duan SY *et al*., 2014). Bone over growth leads gradually to development of exophthalmos and result to complete vision loss by compression of the optic nerve. Nasal breathing and food intake are also influenced as the condition progresses (Dehghan P, Ali Z, 2015; Harut H, Aron B, 2014; Duan SY *et al*., 2014).

Leontiasis Ossea is said to be a rare disease as the overgrowth of bone affects all the cranial bones as well as those of the face, the senses being lost one by one and death finally resulting from cerebral pressure. There is no treatment other than exposing the overgrown bone, and chipping away pieces, or excising entirely where possible (Harut H, Aron B, 2014; Duan SY *et al*., 2014).

EPIDEMIEOLOGY

No epidimeological data available although it is generally assumed that the syndrome is rare. Leontiasis ossea syndromes have been reported in the literature, most of them in subjects ≤18 years old.

ETIOLOGY

The term leontiasis is not used much anymore, since historically it was attributed to a number of different diseases. Although it can be the result of conditions such as syphilis, tumors, and gigantism, the most common disease that causes a non-cancerous overgrowth of the maxilla is craniofacial fibrous dysplasia, a gene mutation that occurs in the early stages of fetal development. Fibrous dysplasia mostly affects children and young adults whose skeletons are still developing.

Leontiasis ossea is divided into true and false leontiasis:

True leontiasis is asymptomatic and false leontiasis is symptomatic. True leontiasis is classified to type1 and type2.

Type1: Injury and infection

Type2: Primary liver disease, Disease of nervous system

CLINICAL MANIFESTATION

A new classification of leontiasis ossea is suggested, by which the condition is divided into *true leontiasis* and *false* (or *symptomatic*) *leontiasis*, True leontiasis is a clinical syndrome caused by two distinct types of disease, whose pathology, however, is related. False leontiasis gives a superficially similar picture but on detailed examination is found to be distinct; it may be caused by a variety of different conditions (Haroyan H *et al*., 2015).

TRUE LEONTIASIS OSSEA FALSE LEONTIASIS OSSEA

TYPE 1: Virchow's type or sclerotic type Paget's disease

TYPE 2: Fibro-dysplastic type which includes: Congenital gigantism

Monostotic fibrous dysplasiaPituitary gigantism

Polyostotic fibrous dysplasia Craniostenosis

Albright's syndrome

Multilocular cystic disease of the jaws

GENERAL SYMPTOMS

Swelling of the left frontal region of the head, loss of vision, Bleeding and difficulty of breathing, fever and may experience pain in eyes

ANATOMICAL CORRELATION WITH SYMPTOMS OF LEONTIASIS OSSEA

Few studies have shown that enlarged parathyroid glands and the patient experienced spontaneous fractures of ribs and changes in the structure of all bones, corresponding to the underlying hyperparathyroidism (changes were revealed by X-ray). The skull and mandible weights were respectively abnormal (high). The cranial vault showed diffuse and irregular proliferation, which produced a grossly in its outer surface. In some areas, bony tended over the suture lines blurring .Eye balls were swollen.

DIAGNOSIS

CLINICAL INVESTIGATION

- Overgrowth of the cranial and facial bone resulting in the expansion of the malar process
- Reducing nasomaxillary angle.
- X-ray

LABORATORY PARAMETERS

- Elevated parathyroid hormone
- Elevated serum concentration
- Elevated ESR

TREATMENT

There is no treatment other than exposing the overgrown bone, and chipping away pieces, or excising entirely where possible. Besides excessive abnormal bone, Immediate postoperative frontal view, Immediate postoperative lateral view enhancing the physical and psychological condition of the patient, surgical treatment is required to prevent the senses being lost one by one and possible death finally resulting from cerebral pressure. Ultimately, early recognition of incipient leontiasis ossea clinically and on imaging is essential to prevent progression to severe disfigurement that can result from prolonged untreated secondary hyperparathyroidism. The importance of early diagnosis and treatment of Leontiasis ossea is very necessary to avoid further complications.

CONCLUSION

Lion face Syndrome is still unknown as it is a rare disease. The syndrome can be treated by adopting a measure such as surgical method. These articles emphasize the relevance of the rare condition, mainly affects the facial bones and bring more awareness among the healthcare sector and in our society.

ACKNOWLEDGEMNET

Nil.

CONFLICT OF INTEREST

None.

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Cite this article:
Sruthi Prasad, Parvathy Nandan, Sruthy Sivan, Haritha k Harikumar, Anjana Sankar A.J, Amina Jabinand Lallu Mariam Jacob .Leontiasis Ossea Syndrome: A Review.*International Journal of Pharmacy & Therapeutics*, 10(3), 2019, 85-87. DOI: http://dx.doi.org/10.21276/ijpt.2019.10.3.6